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Title: A Rare Case of Periapillary Tumor

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Introduction: Gangliocytic paraganglioma is a rare tumor that can affect anywhere in the gastrointestinal tract, though are most frequently found in the second portion of the duodenum. They commonly present with abdominal pain, gastrointestinal bleeding, or rarely with obstructive jaundice. This is a case of an asymptomatic periapillary gangliocytic paraganglioma.

Case: A 45year old male with a history gastroesophageal reflux well controlled on a PPI was referred iron deficiency anemia with no overt bleeding. He underwent an upper endoscopy that revealed a submucosal nodule involving the ampulla (Figure 1). The endoscopic ultrasound subsequently revealed a 2.3x1.5cm hypoechoic heterogeneous mass of the periapillary region (Figure 2). Fine needle aspiration showed a neoplasm with neuroendocrine differentiation. The immunohistochemical stain was strongly positive for synaptophysin and negative chromogranin, S-100, CD-117, and DOG-1. The periapillary mass was resected using endoscopic mucosal resection, with the final pathology consistent with gangliocytic paraganglioma (Figure 3). The patient had further testing with negative urine metanephrines and catecholamines. There was no radiographic evidence of distal metastatic disease.

Discussion: Gangliocytic paraganglioma (GP) is a rare neuroendocrine tumor, with only around 200 cases reported. It consisted on three distinct cellular elements: spindle cells, epithelial cells, and ganglion cells. In spindle-cell predominant tumors, the differential diagnosis includes schwannomas and GIST. When visualized endoscopically, GPs are difficult to distinguish from other submucosal tumors and may appear as a pedunculated or sessile polypoid mass that may be smooth or ulcerated. Endoscopic biopsies are frequently negative since the tumor is submucosal. An EUS is frequently necessary to obtain a fine-needle aspiration as well as evaluate for local spread of the tumor and detect metastatic lymph nodes. The majority of GPs are benign without local or distant spread. However, lymph nodes metastases and distant metastases to other organs have been reported. Treatment is dependent on the size of the tumor and the presence of any metastatic lymph nodes or other metastatic sites. Case reports have shown that overall the prognosis is good but there have been reported deaths from metastatic tumors. In conclusion, we present the case of an incidental periapillary gangliocytic paraganglioma.

*The views expressed are those of the author/presenter and do not reflect the official views or policy of the Department of Defense or its components.

FIGURE 1:



Figure 1: Endoscopic image of the periampullary mass

FIGURE 2:



Figure 2: Endoscopic ultrasound image of the periampullary mass

FIGURE 3:

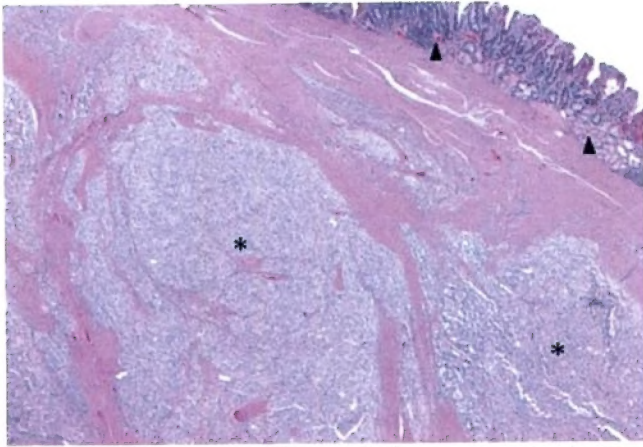


Figure 3: Seven ganglion cells have abundant cytoplasm, round nuclei, and prominent nucleoli (small arrows). Spindle cells are arranged in fascicles (large arrows). Epithelioid endocrine cells are arranged in trabeculae (arrowheads)